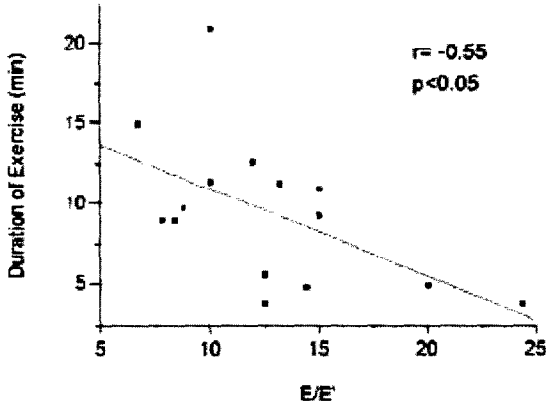
duration.

CONCLUSION: Unlike conventional Doppler indices alone, E/E' ratio correlates inversely with Ex capacity in patient with HCM.



1044-36

Tissue Doppler Derived Index (E/Ea) Correlates With Exercise Capacity in Patients With Hypertrophic Cardiomyopathy

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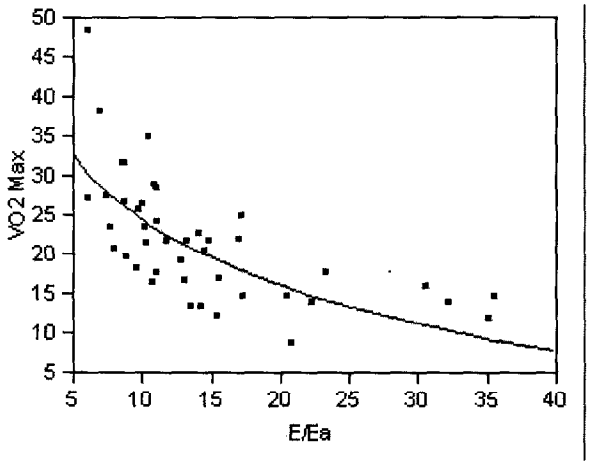
Background: Abnormal LV diastolic function is proposed to contribute to impaired exercise capacity in hypertrophic cardiomyopathy (HCM). In HCM, the ratio of transmitral early filling (E) to mitral annular tissue Doppler (TD) early relaxation velocities (Ea) - E/Ea, is the echo parameter that correlates best with invasively measured LV filling pressures.

Objective: To prospectively determine if E/Ea correlates with exercise capacity in HCM.

Methods: Fifty-nine patients (39 male; mean age 47.7 +/- 17.7yrs) with HCM underwent treadmill stress echo with determination of maximum oxygen consumption (VO2-max). All had mitral inflow and pulmonary venous Doppler, color M-mode and mitral annular TD, immediately before and after exercise.

Results: Traditional indices of LV diastolic function (mitral deceleration time, E/A ratio, pulmonary venous S/D ratio, left atrial area) did not demonstrate significant relationships with VO2-max (mean = 21.8 +/- 8.1ml/Kg/min). LVOT gradient at rest correlated weakly with VO2-max (R = 0.13, p = 0.007). By multivariate analysis, the E/Ea ratio, demonstrated the strongest relationship with VO2-max (r = - 0.7, p < 0.001; VO2-Max = 52.2 - 11.8*Log (E/Ea); SEE = 1.7ml/Kg/min). This was independent of resting LVOT gradient.

Conclusions: In HCM, the TD derived index (E/Ea), an estimate of LV filling pressures correlates significantly with VO2-max, independent of LVOT gradient, suggesting that abnormal diastolic function is an important factor limiting exercise capacity.



1044-38

Early Detection of Fabry Cardiomyopathy by Tissue Doppler Imaging

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Background: Pre-hypertrophy diagnosis of Fabry cardiomyopathy (FC) is actually not obtainable through non-invasive tools.

Methods: We studied 3 groups of patients (pts): 10 pts with mutations for Fabry disease and left ventricular hypertrophy (LVH), 10 mutation positive pts without LVH and 10 healthy relatives. All pts with LVH and six Fabry pts without LVH presenting with ventricular arrhythmias underwent cardiac MRI and biventricular endomyocardial biopsy to assess cardiac involvement. In all pts two-dimensional echocardiogram with tissue Doppler analysis in the pulse-doppler mode was performed: systolic (Sa), early diastolic (Ea), and late diastolic (Aa) velocities were measured, and the Ea/Aa ratio and the E/Ea parameter computed at both corners of the mitral annulus.

Results: Histology and electron microscopy showed glycospyngolipids deposits in all cases. All mutation-positive pts had significant reduction of Sa, Ea and Aa velocities at both corners of the mitral annulus compared with controls. Ea/Aa ratio was lower and E/Ea ratio higher in mutation-positive pts than in controls (p<0.001). Pts with LVH showed lower contraction and relaxation TD velocities, lower Ea/Aa ratio and higher E/Ea ratio in comparison with mutation-positive pts with no LVH (p<0.001).

Conclusions: FC presents reduced myocardial contraction and relaxation TD velocities appearing before the development of LVH. TDI can provide a pre-clinical diagnosis of FC allowing early institution of enzyme replacement therapy.

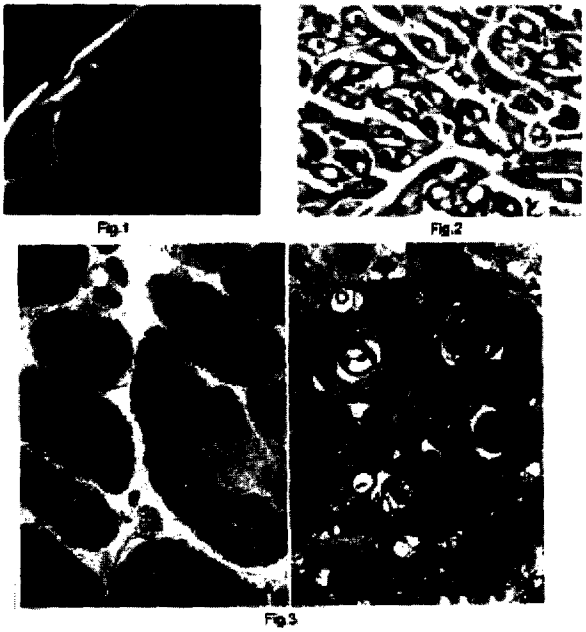

Fig.1 Fig.2

Fig.3

Fig. 1. Cardiac magnetic resonance of a mutation-positive patient without LVH showing normal ventricular wall thickness and no abnormalities of myocardial signal intensity.

Fig. 2. Left ventricular endomyocardial biopsy of the same patient showing mildly enlarged myocardiocytes containing perinuclear vacuoles. Hematoxylin and Eosin, 250X.

Fig. 3. Transmission electron micrographs of left ventricular endomyocardial biopsy of the same patient showing at low (a) and high (b) magnification the perinuclear vacuoles to consist of single membrane-bound vesicles containing concentric, lamellar, electron-dense figures, typical of glycolipid storage disease. (a) 1250 X, scale bar=10 micron. (b) 11000 X, scale bar=1 micron.

1044-39

Reduced Longitudinal Strain Rate in Patients With Cardiac Amyloid Despite Preserved Fractional Shortening Equals That of Dilated Cardiomyopathy

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Background: A significant difference in strain rate for patients with cardiac amyloid with a history of CHF vs. no history of CHF has previously been observed. However, the extent of this abnormality and its dependence on fractional shortening (FS) is not known. We sought to quantify this abnormality by comparing cardiac amyloid to two groups: patients with severely depressed FS (dilated cardiomyopthy(CMY)) and normal FS (normal subjects).

Methods: 99 patients with biopsy proven amyloid - 53 patients with no history of CHF and 46 with a history of CHF (of those with CHF, 18 had a FS > 30%, and 28 had a FS < 30%), 14 patients with CMY (nonischemic, mean FS 20%), and 19 normal subjects were recruited. We previously found that peak systolic strain rate (PSSR) at the basal septum of the 4-chamber view can reflect global contractility when segmental differences are not present; therefore, all measurements were taken at this site.

Results: A significant difference in PSSR exists between normals and all other groups (table). This was true even for patients with cardiac amyloid who had no history of CHF and normal FS; in fact, we found no difference between this group and patients with CMY ($p > 0.05$).

Conclusions: PSSR, a marker of longitudinal systolic function, is significantly impaired in cardiac amyloid even in the absence of CHF and despite normal FS. This impairment,